NEWER METHODS IN THE DIAGNOSIS OF CONGENITAL CARDIAC ANOMALIES*

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DURING the last decade, studies of congenital heart anomalies have received great impetus, since corrective surgical procedures have been proposed for the treatment of the most common forms observed: Patent ductus arteriosus, coarctation of the aorta, pulmonic stenosis isolated or combined with other malformations, interauricular and interventricular septal defects. The selection of suitable cases for surgery has therefore become a clinical problem of considerable interest to practitioners who, in order to discharge properly their responsibility, should be well informed of new developments in methods of diagnosis.

In this lecture, I shall attempt to review our present day knowledge of two methods which have received widespread attention: Cardiac catheterization and Angiocardiography.

DISCUSSION OF METHODS

1. *Cardiac Catheterization*

The introduction of a long radio-opaque, modified ureteral catheter into the venous system under fluoroscopic control offers a means of sampling blood in the superior and inferior vena cava, in the right auricle, the right ventricle, the pulmonary artery and its branches. In a subject maintained in a steady state, variations in the oxygen content of mixed venous blood samples taken in rapid succession in these chambers and vessels are small. Abnormal communications between one of these right-sided structures and its adjacent counterpart on the left side of the heart, i.e., the left auricle, the left ventricle or the aorta, result

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in contamination of mixed venous blood returning from the tissues by well-oxygenated blood returning from the lungs, because the pressure conditions prevailing in the circulation favor blood shunts from left to right. Therefore, the approximate location of shunts can be determined by gas analysis of blood samples drawn through the catheter as its tip is placed in various right-sided cardiovascular structures. Moreover, the presence of an abnormal communication may be revealed by the unusual routes eventually taken by the tip of the exploring catheter as it is introduced into the heart.

Collection of arterial blood, through an indwelling needle, and determination of the oxygen content and saturation in the samples drawn may help also in recognizing contamination of well-oxygenated blood by mixed venous blood. Such contamination occurs when the dynamic conditions prevailing in the heart in the presence of a cardiac malformation, favor a shunt directed from the right cardiac cavities or the pulmonary artery into the left cardiac cavities or the aorta.

With simultaneous determinations of the oxygen intake into the lungs, and of the oxygen contents in the right heart and pulmonary artery, and of the oxygen content in a systemic artery, the calculation of blood flow in the systemic and in the pulmonary circuits and of blood shunts through abnormal communications, becomes a matter of applying simple formulas.

The technique of right heart catheterization provides also a means of recording blood pressures. The patterns of the curves and the range of normal pressure variations in the right and left auricle, the right and left ventricle, the pulmonary artery and the aorta, and in a peripheral artery, when tracings are obtained through an indwelling needle, are well defined. Hence, these blood pressure recordings may serve two purposes: 1) Identification of the cardiac cavities or large vessels in which the tip of the catheter is placed; 2) study of the effects of cardiac malformations upon the dynamics of the circulation.

Several conditions must be met if the results of cardiac catheterizations are to be considered valid and justifiable of an interpretation: 1) Blood samplings and pressure recordings must be made under fluoroscopic control. 2) Blood samplings, in the same vessels and cavities, must be multiplied and obtained in rapid succession. 3) A steady state of the respiration and circulation must be maintained during the whole period of samplings.

In young children, a general anesthetic may be required in order to maintain a steady state during the procedure. Multiple determinations of the oxygen saturation in blood samples may be facilitated and may even be instantaneously observed by the use of an oxymeter of special design. The blood pressure recordings must be made immediately visible, by means which depend upon the type of recorder in use.

Even though favorable technical conditions have been obtained, the interpretation of the data requires good judgment. The same differences in the oxygen content of the blood in two heart chambers may be due to different abnormalities. For instance, an increase in oxygen content in the right ventricle, as compared to the right auricle, in the presence of a patent ductus arteriosus, may be either due to pulmonary regurgitation or an associated interventricular septal defect.

The calculation of systemic or pulmonic flows and of shunts must be considered at best as an approximation, because it is not always possible to ascertain whether a sample obtained near, or even away from the main stream of a shunt, is adequately mixed with the rest of the blood flowing in the cardiac cavity or large vessel.

Most *complications* observed during and after cardiac catheterizations in the course of study of congenital cardiac anomalies, are of a minor character. Air emboli introduced into the catheter are potentially dangerous in cases where, hemodynamic conditions favoring right to left shunts, small air bubbles may be thrown into the systemic circulations. Precautions should therefore be taken to avoid introduction of bubbles into the catheter. A traumatic lesion of the endocardium with secondary thrombosis is another potential danger to be reckoned with. Although their frequent occurrence in the dog has been well documented, these lesions, to my knowledge, have been demonstrated in one single instance in humans, although they have been looked for by many investigators, when the occasion of doing so presented itself. Acute bouts of anoxia, following catheterization of a markedly stenotic pulmonary valve associated with an interauricular communication have been observed. Proper care should therefore be exercised in avoiding a prolonged obstruction of such a stenotic area especially when cyanosis develops, or its degree increases during manipulation of the catheter. Development of rhythm disturbances is by far the most important complication to consider. Since continuous electrocardiographic control has become a routine part of the exploration of the cardiac cavities and

of the thoracic great vessels, it has been found that ectopic beats, ectopic rhythms and tachycardias are not exceptional, and that conduction disturbances may develop, although much less frequently. The arrhythmias are more apt to occur when the tip of the catheter is in the infundibular area of the right ventricle, or introduced in the left ventricle through a septal defect. Save in exceptional instances they cease immediately upon displacement of the catheter, and have no clinical repercussion. But in the presence of prolonged bursts of ventricular tachycardia, one should fear a marked drop in systemic pressure which can in cases with cyanosis bring about a considerable increase in arterial anoxia and even more fear the development of ventricular fibrillation. Since the mortality rate attributable to this technique can be estimated from available figures at about two tenths of one per cent, it would appear that the risk is small indeed. It is nonetheless one that has to be taken into account before deciding to proceed with the technique.

From the preceding remarks, it is obvious that cardiac catheterization should only be undertaken by a well-trained team, in a well organized laboratory. The reliability of the results and the safety of the method depends upon meeting both these requirements.

2. *Angiocardiography*

Angiocardiography, as is well known, consists in the x-ray study of the cardiac chambers and thoracic great vessels during opacification by an intravenously injected radio-opaque solution. This method has been widely used in recent years for the diagnosis of various types of cardiac congenital malformations. With x-ray films of the chest taken in rapid sequence, it is possible to delineate the contour of the four cardiac chambers and of the large vessels, to visualize details of structures otherwise invisible, such as the valvular areas, and also to follow the order in which the cardiac cavities and large vessels are filled by the blood conveying this radio-opaque substance. Hence, details of abnormal anatomy, and unusual pathways taken by the circulating blood, may be revealed in the course of study of congenital heart anomalies, especially since x-ray exposure may be obtained now at the rate of several films each second in two different projections.

Introduction into the circulation of a radio-opaque substance, however, is not without some *risk*. In a very well documented paper, Dotter and Jackson have reported the result of a survey which revealed that

out of a total of nearly 7,000 angiocardiographic studies in 182 hospitals, twenty-six deaths had occurred which could be attributed to the injection of the radio-opaque substance. All but two of these deaths were observed in subjects with congenital heart diseases, most of them belonging to the cyanotic group. These figures indicate that angiocardiography is not to be undertaken lightly, and without sufficient indication in this type of patients.

Before analyzing the nature of the information supplied by both these new methods in the most prevalent forms of congenital cardiac anomalies, it must be emphasized that their results must be compared with, and often interpreted in the light of observations obtained by careful and exhaustive physical and x-ray examinations. Neither of these newer methods can be considered as a substitute for tried and sound clinical studies. Furthermore, coöperation between clinicians, roentgenologists and clinical physiologists is essential. It would be even better, in my estimation, if the clinician who is responsible for therapeutic decisions, could himself master these techniques, or at least be able to interpret their results and recognize their inadequacies.

Until the advent of these methods of exploration, advances in the knowledge of congenital heart disease has been relatively slow, and based essentially upon correlation between clinical observations and post-mortem examinations. Although the admirable achievements of Maud Abbott, J. B. Brown and Helen Taussig cannot be too highly praised, the anatomo-clinical method has in some instances created distorted views and false impressions. Cardiac catheterization and angiocardiography made possible a study of anatomic anomalies and dynamic processes in the living. Not only have they helped to discover that the incidence of certain malformations is greater than suspected, but they have added new significance to the interpretation of physical and x-ray signs, which to a great extent are expressions of these processes. Briefly, the introduction of these methods in the study of congenital heart disease has resulted in a sharpening of the diagnostic acumen of clinicians engaged in the study of these fascinating and often baffling cardiac anomalies.

Discussion of Results

1. *Non-Cyanotic Congenital Heart Anomalies*

We shall first examine the value and significance of data obtained

by cardiac catheterization and angiocardiography, in the most prevalent forms of the non-cyanotic types of congenital heart disease: Coarctation of the aorta; patent ductus arteriosus; interventricular and interauricular septal defects and finally, pulmonic stenosis, either isolated, or associated with a septal defect through which there is no reversal of the natural direction of shunt.

Coarctation of the Aorta: In coarctation of the aorta of the so-called adult type, a stenosis of a part of the aorta distal to the left subclavian artery is associated with hypertension in the upper extremities, and a lower blood pressure in the lower extremities, where the circulation is maintained as a result of the development of large anastomoses bridging the stenotic area.

Cardiac catheterization has no place in the study of such an anomaly, whereas the information obtained by angiocardiography can be considered as essential, and sometimes decisive in deciding the surgical approach.

The radio-opaque substance once it reaches the involved area, precisely delineates the nature and extent of the lesions. The left anterior oblique is the position of choice for the study.

As a rule one can easily recognize left ventricular enlargement; an ascending aorta slightly or markedly dilated; an innominate artery and a left subclavian artery greatly dilated; and a transverse aortic arch narrow and long. The site of the coarctation is identified as a short stub extending downwards and posteriorly beyond the subclavian artery, and appearing like the end of a sausage. The descending aorta usually dilated, overlaps slightly the stub, and as a rule, the opening between the two segments is not demonstrable readily. The timing of opacification of the descending aorta is *not* related to the size of the opening, but to the character of the collateral circulation. A pre-operative study of the site, length of the narrowed area, origin of collaterals, and of other branches of the arch of the aorta, permits as a rule the planning of the type of reparative surgery to be employed, especially whether a vessel graft should be available.

Rarely does angiocardiography fail to afford adequate demonstration of the abnormality. If it should fail, one may resort to retrograde aortography which consists in cannulating a carotid artery, or passing a long and narrow polyethylene tube up the aortic arch through a brachial artery, and injecting the contrast substance through the cannula or tube.

Potential dangers attendant to this type of exploration should limit its use to the exceptional case.

Patent Ductus Arteriosus: The persistent patency of a ductus arteriosus with continued communication between the aorta and the pulmonary artery results in a substantial volume of blood being shunted from the former into the latter, in accord with the large pressure difference existing between both vessels during the entire cardiac cycle. Hence the work of both ventricles, especially the left, and the pulmonary blood flow, are both increased. The large pulmonary blood flow together with increased pressure at the site of the shunt combine to cause the dilatation of the main pulmonary artery and of its major branches.

In most instances the clinical diagnosis is obvious, and the decision regarding surgical exploration arrived at without recourse to either cardiac catheterization or angiocardiography. However, in some cases the diagnosis may remain in doubt. This happens particularly in cases where a systolic murmur is present at the base with a collapsing type of pulmonary circulation as viewed by fluoroscopy; or when an additional systolic murmur is heard in the precordial area, or when the continuous classical murmur has its maximum lower than the second left interspace, and finally, when a diastolic murmur of pulmonary regurgitation complicates the picture.

Among the helpful information supplied by *cardiac catheterization,* one must list the direct demonstration of a patent ductus arteriosus by insertion of the tip of the catheter into this channel by way of the left pulmonary artery near its origin and the emergence of the tip in the descending thoracic aorta. Thus in left anterior oblique the opaque catheter appears as a cord subtending the arch of the aorta. An indirect demonstration of the shunt of well-oxygenated blood from the aorta to the pulmonary artery is provided by the finding of a considerable increase in the oxygen content in the latter vessel as compared to the oxygen content of the right ventricle and auricle.

Whereas analysis of the oxygen content in the successive blood samples obtained from the two right chambers and from the pulmonary artery cannot differentiate an aortic septal defect from a patent ductus arteriosus, the direct demonstration of the course followed by the catheter through the patent ductus is pathognomonic. However, the coarser character of the murmurs and thrills, their lower locations, a history of subacute endocarditis, are as a rule clinically suggestive of the

diagnosis of an aortic septal defect, a much rarer condition.

In patent ductus arteriosus, calculations indicate that as a result of the shunt, the pulmonary blood flow may reach two to four times that of the systemic blood flow. The blood pressures in the pulmonary artery may be normal, or sometimes considerably elevated, but there appears to be no absolute relationship between the height of pressure, the size of the shunt or the age of the subject. However, it must be borne in mind that as a result of prolonged or considerable increase in pulmonary blood flow atheromatous changes occur in pulmonary vessels, causing an increase in pulmonary vascular resistance which is reflected in a rise of pulmonary artery pressure, while as a result of this increased resistance, the magnitude of the aorta-pulmonary shunt diminishes. In some instances this process may lead to reversal of flow, and late cyanosis. In the presence of a complicating functional pulmonary valve incompetence which during diastole causes regurgitation of blood with a high oxygen content into the right ventricle, the average oxygen content in this cavity increases significantly above that of the right auricle. Under such conditions, the differential diagnosis on the bases of blood gas determinations may become difficult with 1) a high interventricular septal with persistence of laminar flow in the pulmonary artery, 2) a high ventricular septal defect complicated by aortic insufficiency, and 3) a combination of patent ductus arteriosus with an interventricular septal defect. Confusion, then, may result from the analysis of data obtained by cardiac catheterization, unless a very keen analysis is made of the character, location and propagation of murmurs present.

Angiocardiography is seldom used as a diagnostic procedure in patent ductus arteriosus. Several signs have been claimed to be of relative or absolute diagnostic value, i.e., 1) a localized dilatation of *the aorta* distal to the origin of the left subclavian artery, 2) enlargement of the pulmonary artery, with elevation of its left main branch; 3) reopacification of the left pulmonary artery at the time of the aortic opacification; 4) actual opacification of the patent ductus arteriosus.

Although the presence of the above mentioned localized dilatation of the aorta may be considered good presumptive evidence of a patent ductus arteriosus, it may occur in other conditions, and is often absent in proven cases. As for the actual opacification of the patent ductus arteriosus, it is exceptional unless the contrast medium is introduced by the method of retrograde aortography.

Interventricular septal defect: Two distinct locations of interventricular septal defects correspond to two distinct clinical and physiological pictures: 1) A simple perforation through the muscular part of the septum associated with characteristic discrete findings of the classical "maladie de Roger," with small volume of blood shunted from left to right, and a slight load placed on the heart which remains normal in size and shape. 2) A high septal defect located just below the semi-lunar valves which is associated with a large shunt, pulmonary hypertension, dilatation of the pulmonary artery, and hypertrophy of both ventricles.

Cardiac catheterization and *angiocardiography* are unnecessary in the first type, although the presence of the shunt may be demonstrated by gas analysis of blood samples, if it is of sufficient magnitude. In the second type, *cardiac catheterization* is of great help in demonstrating an increase in oxygen content in the right ventricle, particularly in the samples obtained in the infundibular area and the presence of pulmonary hypertension. As discussed previously the exclusion of the diagnosis of patent ductus arteriosus with pulmonary incompetence may present, even after catheterization, some difficulty.

Angiocardiography is of little use in this type of interventricular communication. If performed, it may occasionally show a persistent filling of the right ventricle and of the dilated pulmonary artery during left ventricular and aortic opacification.

Interauricular septal defect: The persistence of an ostium primum or secundum, or of a patent foramen ovale, allows blood to pass from one atrium to the other, the direction of the shunt depending upon the pressure relationship between both atrial cavities. Uusually, unless there is considerable dilatation of the right atrium, the shunt is predominantly from the left to right, as the pressures in the left auricle are normally higher than in the right, causing therefore an increase in pulmonary blood flow, and consequently, right ventricular enlargement and often dilatation of the pulmonary artery.

Cardiac catheterization may help demonstrate the presence of this defect either directly or indirectly: (a) Introduction of the tip of the catheter through the defect in the left auricle and into one of the pulmonary veins is easily recognized and usually, but not always, achievable. (b) The demonstration of a left to right shunt is based on the observations of a higher oxygen content in the blood sampled from the right atrium than in the mean value of blood oxygen from superior and inferior

venae cavae. Occasionally, if the tip of the catheter is at the time of blood sampling immediately in the path of the stream of highly oxygenated blood spurting from the left auricle, the demonstration of fully oxygenated blood becomes pathognomonic.

It should also be emphasized that in interauricular septal defects laminar blood flow may persist in the ventricle and even in the pulmonary artery causing variations in oxygen content in these chambers, especially when the shunt is large. This makes multiple sampling in this cavity and vessel essential in order to avoid confusion.

The pulmonary blood flow is sometimes greatly increased, reaching two to three times that in the systemic circulation, but may be also only slightly increased. Pulmonary arterial pressure which is as a rule normal or nearly normal, may be elevated occasionally and for no known reason.

The demonstration of an interatrial defect is of considerable diagnostic value in differentiating this malformation from others also associated with dilatation of pulmonary artery and with evidence of a large or even normal pulmonary blood flow. To the group with a large blood flow belongs the high ventricular septal defect, associated usually with high pulmonary arterial pressure; to the group with a normal, or nearly normal pulmonary blood flow, belong a variety of anomalies that we shall discuss shortly: Pulmonic stenosis of mild degree, Idiopathic dilatation of the pulmonary artery, and "Primary" pulmonary hypertension with pulmonary valve insufficiency.

As for the differentiation between an interauricular septal defect and direct drainage of a pulmonary vein into the right atrium, the solution depends upon the direction taken by the tip of the catheter, which in some instances may be revealing.

If and when *angiocardiography* is performed, x-ray exposure is preferable in the frontal view in adults, and in the left anterior oblique view in children. The films demonstrate a large right auricle, right ventricle and pulmonary artery, with engorged branches of the pulmonary artery, while the left auricular and ventricular cavities and the aorta are of normal size. Opacification of the right chambers and pulmonary artery persists after the left heart has been filled, but great caution should be exercised in interpreting this finding as a slow injection with pooling in the hepatic vein and jugular veins of some of the contrast medium may result in the same observation. In children the instantaneous spreading of the contrast medium from the right to the left atrium may be demon-

strated directly. This temporary reversal of flow through the defect is probably due to the temporary pressure increase in the right auricle, following the rapid injection of a fairly large quantity of liquid which has a small distance to run from the arm to the right auricle. Dotter and Steinberg estimate that with the multiple exposure technique this sign is present in about one-third of the cases only.

Pulmonic Stenosis: The problem of pulmonic stenosis not associated with dextroposition of the aorta, may be said to have been greatly clarified since congenital heart anomalies have been systematically studied by cardiac catheterization.

A. *Simple valvular stenosis and infundibular stenosis with closed septa* have been demonstrated in many instances. They are of two types, mild and severe, both characterized by pulmonary artery dilatation with normal pulmonary vascular markings. In the mild form, usually symptomless, there is no demonstrable right ventricular enlargement, whereas in the severe form, cyanosis of the peripheral type, i.e., not associated with arterial blood anoxia, breathlessness, and sometimes syncope and angina pectoris, may be observed. Evidences of right ventricular hypertrophy and dilatation with or without right ventricular failure, become more striking according to the degree of stenosis and the stage of evolution.

Cardiac catheterization provides characteristic findings namely a normal or low pulmonary arterial pressure associated with a moderate or considerable systolic hypertension in the right ventricle. The exact location at which the systolic pressure suddenly rises, when the tip of the catheter is withdrawn from the pulmonary artery into the right ventricle under fluoroscopic control, will help in differentiating valvular from infundibular stenosis. This information may be of considerable value, for a different type of valvulotomy may be used for the relief of each of these malformations. The oxygen concentration in all the samples removed from the right side of the heart and pulmonary artery, being nearly identical, closure of the septa is thus demonstrated.

In the mild form of pulmonic stenosis, the most common error made in differential diagnosis is that of suspecting an atrial septal defect with small shunt or a "maladie de Roger." In some cases with a mild degree of stenosis associated with an aneurysmal dilatation of the pulmonary artery, the only other diagnosis to be discussed is that of idiopathic dilatation of the pulmonary artery, where the systolic pressure in the right ventricle is not elevated, although the systolic pressure in the pul-

monary artery may be somewhat lower. In the severe forms of stenosis, confusion with any other conditions is unlikely as soon as it is demonstrated that the cyanosis is peripheral and therefore not associated with a reduction in the arterial blood oxygen saturation.

Angiocardiography is of little help, besides revealing the contours of the pulmonary artery and its main branches. Rarely does it demonstrate a narrow and irregular channel, either at the valve area or in the infundibular area. Great caution must be exercised in the interpretation of such details and cardiac catheterization should be performed if the diagnosis is in doubt.

B. *Pulmonic stenosis associated with septal defects but without reversal of blood flow* is also not an exceptional anomaly. The clinical signs of either the stenosis or the septal defect may dominate the course and the diagnosis may be puzzling.

Cardiac catheterization constitutes the keystone of the diagnosis in revealing both types of defects: 1) By supplying direct evidence of pulmonic stenosis derived from the blood pressure tracings and 2) a direct or an indirect demonstration of an associated septal defect by observations which have been previously commented upon.

Before we start a discussion of the use of these new methods in the cyanotic group of cardiac malformations, we may briefly summarize their essential indications in the acyanotic type:

1. *Cardiac catheterization* is especally important in subjects with enlargement or dilatation of the pulmonary artery, whenever the character of the murmur and thrill and other physical signs leave some doubt as to the exact nature of the defect, and when the surgical decision depends upon the accuracy of the diagnosis.

2. *Angiocardiographic studies* are indispensable in the preoperative study of coarctation of the aorta.

2. Cyanotic Types of Congenital Cardiac Anomalies

From the point of view of incidence as well as that of surgical interest, there are two major types of congenital heart anomalies, accompanied by cyanosis from early life on, namely (a) severe pulmonary stenosis with septal defect and reversal of shunt from right to left, and (b) severe pulmonary stenosis with a dextroposed (overriding) aorta which saddles a high interventricular septal defect.

Pulmonic stenosis with septal defects and reversal of shunt: Severe

pulmonic stenosis in the first of these anomalies causes 1) a reduction in pulmonary blood flow, but nonetheless a post-stenotic dilatation of the pulmonary artery is often present; 2) a considerable systolic hypertension in the right ventricle with hypertrophy of the wall of this chamber; 3) a dilatation of both right ventricle and right auricle with increase in the diastolic filling pressure. The later pressure rise results in a reversal of blood flow through the associated septal defect, usually interauricular or the patent foramen ovale.

As a rule, the clinical diagnosis is based on a history of rapid progression of a cyanosis which may have been mild at birth, and its association with a dilatation of the pulmonary artery. Since pulmonary valvulotomy rather than a Blalock anastomosis should be performed in this malformation, cardiac catheterization and angiocardiography are of great value in discarding the diagnosis of Tetralogy of Fallot. It should, however, be emphasized that the correct diagnosis may be strongly suspected on clinical grounds.

By *cardiac catheterization*, direct evidence may be attained of the presence of a pulmonic stenosis by pressure tracings. The systolic hypertension in the right ventricle is always considerable, reaching sometimes twice that in the peripheral systemic artery. Direct evidence of an interauricular communication is obtained, if the catheter tip may be slipped from the right auricle into the left auricle. It is notable that when there is reversal of flow through the interauricular communication, the oxgyen content in the right auricle, right ventricle and the pulmonary blood samples are identical.

Angiocardiography reveals (a) an early shunt of blood from the right auricle to the left auricle by simultaneous filling of both these cavities, (b) an early opacification of the left ventricle and aorta, (c) a minimal opacification of the right ventricle and of the dilated pulmonary artery, (d) a late reopacification of the left auricle. All these findings are of great diagnostic value. Occasionally, the character and location of the stenosis may also be revealed.

In exceptional cases, a severe pulmonic stenosis is associated with an interventricular septal defect with reversal of shunt. This diagnosis will be considered, in the discussion of the classic Tetralogy of Fallot, where in addition dextroposition of the aorta is present.

Pulmonic stenosis with dextroposition of the aorta (Tetralogy of Fallot). Combination of these two defects, pulmonic stenosis and dextro-

position (overriding) of the aorta, underlies the most common types of congential cardiac malformations with cyanosis present since birth.

The overriding of the aorta implies of necessity a high interventricular septal defect; the semi-annular form of this defect is distinct in general from the annular form of the isolated high septal defect previously depicted. The right ventricular hypertrophy constitutes the fourth element of the classical Tetralogy of Fallot, under which title this group of anomalies are best known. The degree of ventricular hypertrophy which is most likely dependent upon the degree of pulmonic stenosis and of dextroposition of the aorta, is variable, but never reaches that which is seen in some cases of isolated severe pulmonic stenosis. The anatomic and physiologic variants of this type of anomaly are numerous. They are dominated by 1) the degree of stenosis which may range from a slight narrowing to complete atresia and 2) by the degree of overriding of the aorta which may vary from slight to extreme. Dynamically each ventricle may maintain its identity, or both may function as a single ventricle. The distinction between specific types such as classic Tetralogy of Fallot, pulmonary atresia, pseudotruncus arteriosus, extreme dextroposition with common truncus, where some degree of oxygenation of blood in the lungs is maintained, mainly by the markedly dilated bronchial arteries, is not only of academic interest, but also of practical value, since some clinicians consider that pulmonic atresia is a contraindication to a Blalock-Taussig type of operation, and since all agree that the operation is not physiologically feasible in Common Truncus. The respective amount of blood flowing through the pulmonary artery and its branches, and through the aorta, is determined primarily by the respective resistances to flow at the site of the stenosis and in the entire aortic system. As a rule the flow through the main pulmonary artery is considerably reduced, or even absent, as in atresia—in mild form of stenosis it may occasionally be as large as in the aorta. It also stands to reason that the degree of arterial blood oxygen unsaturation and therefore, of cyanosis, is determined by the amount of mixed venous blood shunting directly into the overriding aorta.

The clinical diagnosis is as a rule simple, when the physical signs and x-ray findings are according to the classical description. The exact diagnosis may, however, require special investigations, in particular when (a) the cyanosis is only mild, (b) a post-stenotic dilatation of the pul-

monary artery is present, (c) the reduction in pulmonary blood flow is small, (d) pulmonary atresia with considerable collateral circulation is present. These studies are primarily directed at demonstrating preoperatively whether palliative surgery is indicated and if so, to give advance information concerning choice and modality of techniques to be employed.

In the course of *cardiac catheterization*, the probing tip of the catheter as it progresses into the right ventricle may directly demonstrate the overriding aorta. In such instances, it is seen progressing into a right or left arch, and descending aorta. Pulmonary stenosis may also be demonstrated, if the catheter is introduced into the pulmonary artery by recording pressure tracings. Whereas the pressures are low in the pulmonary artery, the systolic pressure is elevated in the right ventricle, to a varying degree and sometimes attains the systolic pressure level measured in the aorta, or in the systemic artery. The oxygen contents in the blood sampled from the right auricle and the right ventricle and even the pulmonary artery, may be the same when the amount of shunt from the left ventricle to the infundibular area is insignificant. Not uncommonly, however, evidence of some small degree of left ventricular blood shunting appears in this latter region of the right ventricle. Samples of blood collected in the aorta or in a peripheral arterial blood, although more or less unsaturated with oxygen, have always a higher saturation than the right ventricular blood. This evidence helps to exclude the diagnosis of total transposition of the aorta, where the vessel originating from the right ventricle, has the same composition as the right ventricular mixed venous blood. In cases of atresia, or pseudo-truncus arteriosus, where the tip of the catheter cannot be introduced in the pulmonary artery, the degree of blood oxygen unsaturation in the aorta or a peripheral artery, is always considerable, but less so than in the right ventricle. Moreover, it must be strongly emphasized that failure to catheterize the pulmonary artery is no proof of the presence of an atresia. Similarly, failure to introduce the tip of the catheter into the aorta, cannot be given as an evidence that there is little or no dextroposition of this vessel.

Angiocardiography demonstrates superbly the essential defect by simultaneous opacification of the aorta and pulmonary artery, at the time of right ventricular opacification. This picture is almost pathognomonic and quite distinct from that observed in a single ventricle,

where the entire ventricular cavity and both large vessels are opacified simultaneously. The site of the stenosis may sometimes be difficult to demonstrate, but a poor pulmonary vascular filling is a sign of a reduction in the blood flow in the pulmonary artery. In the case of atresia, aortic opacification takes place alone without opacification of the pulmonary vessels. It is exceptional in this malformation to be able to visualize clearly the bronchial vessels, as they branch off the aorta.

Dextroposition of the aorta without pulmonary stenosis: Dextroposition of the aorta may sometimes develop without concomitant pulmonary stenosis. This anomaly has been much discussed in recent years under the heading of Eisenmenger complex. From the experience gained by the use of the new techniques of diagnosis it would seem that the incidence and the importance of this syndrome has been greatly overrated.

In its typical form the complex is characterized by the shunting of a large amount of left ventricular well-oxygenated blood, through an interventricular septal defect, into the pulmonary vascular system where normally resistance to flow is lower than in the aortic system. This results in a large increase in pulmonary blood flow, and the pulmonary artery becomes markedly dilated. The amount of mixed venous blood flowing directly in the overriding aorta is usually small, and therefore, cyanosis is minimal or absent, although the arterial blood oxygen saturation is always slightly reduced. However, under hemodynamic conditions not always well defined, the resistance in the pulmonary vascular bed may increase to such an extent that left to right shunt is progressively reduced, as the pulmonary arterial pressure increases, and mixed venous blood shunts from the right ventricle to the dextroposed aorta. It is in such instances that the dilatation of the pulmonary artery associated with a reduction in the pulmonary blood flow and with increasing evidence of contamination of the arterial blood by mixed venous blood creates a picture which simulates (a) a Tetralogy of Fallot with mild pulmonic stenosis and post-stenotic dilatation of the pulmonary artery; (b) a high interventricular septal defect with considerable pulmonary hypertension and reversal of shunt; (c) an idiopathic pulmonary hypertension with reversal of blood flow through a patent foramen ovale.

Catheterization of the heart is of great diagnostic value if it simultaneously demonstrates: dextroposition of the aorta, absence of pulmonic stenosis, and identical systolic pressures in the pulmonary artery,

the right ventricle and the aorta.

The *angiocardiographic findings* of a simultaneous opacification of the pulmonary artery and aorta, although suggestive, is not diagnostic, as it may be present in all the anomalies under discussion.

To summarize the indications of cardiac catheterization and angiocardiography, in the most prevalent forms of cardiac congenital anomalies of the cyanotic group, it may be stated that both are of great diagnostic value, especially the latter which according to Dotter and Steinberg should be performed routinely in all cases considered for surgery.

Conclusion

Since the introduction of cardiac catheterization and angiocardiography in the study of cardiac congenital anomalies, additional diagnostic information has been gained. Many physical and roentgenologic signs have acquired new meaning and significance, so much so that clinicians familiar with these new techniques feel that they have become unnecessary in many instances. In this presentation an attempt has been made to formulate clear-cut indications for the use of either or both methods. However, because their use entails a minimal but definite risk, especially in the hands of the neophyte and of the daring, a note of caution has been sounded and it has been stressed that good judgment should always be exercised and instantaneous electrocardiographic control should always be available in the course of cardiac catheterization. A further conclusion should emerge from this discussion, i.e., that only those cases should be studied where the diagnosis after careful and expert clinical examination, is still in doubt, and if a better documented diagnosis may be of aid in reaching a therapeutic decision. To the credit of these new methods stand foremost 1) a truer picture of the clinical incidence of some defects which are justifiable of surgery and 2) the considerable progress which has been achieved in the understanding of the physiopathology of the various defects, and of their influence upon the dynamics of the circulation.

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